Original Article

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Pediatric Ureteroceles:
Diagnosis, Management and Treatment Options

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Abstract

Objective: The aim of the study was to evaluate clinical characteristics of ureteroceles particularly for diagnostic and treatment challenges.

Methods: Data about patients treated for ureterocele in the two hospital clinics during 1996-2009 are retrospectively evaluated.

Findings: There were 12 girls and 7 boys. Symptomatic urinary tract infection was found in twelve cases. Ureterocele was associated with duplex systems in eleven cases. Vesicoureteral reflux was detected in 4 patients. Bladder diverticulum complicated with ureterocele in 1 patient. Ultrasonography diagnosed ureterocele in 12 patients. Renal scarring was detected in 6 patients at the side of ureterocele. Fifteen patients showed varying degrees of hydro-ureteronephrosis. Surgical therapy included upper pole nephrectomy in 3 cases. Bladder level reconstruction was performed in 11 cases. Five patients were treated only by endoscopic incision. In the follow up period 4 patients showed long term urinary tract infections whereas 3 of them were treated endoscopically. Postoperative reflux was still present in two patients who were treated by endoscopic incision.

Conclusion: Ureterocele diagnosis and treatment show challenges. Urinary tract infection is important marker for urinary system evaluation. Preoperative management generally depends on a combination of diagnostic methods. Endoscopic incision needs serious follow up for postoperative problems.

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Introduction

Although medical progress including new imaging modalities and technical support has been widely gained, controversy still continues in management of ureteroceles regarding classification, diagnosis, and treatment. The incidence of ureterocele is variable with the highest rate of 1 in 500 and it is generally found in females with duplex system association

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(95%)[1,2]. Though there is a female dominance, the nature of the anomaly is more complex in boys. No one theory explains the etiology for all types of ureteroceles[3-5]. Beginning from the prenatal stage, this anomaly causes different clinical presentations, such as antenatal hydronephrosis, vesicoureteral reflux (VUR), urinary tract infection (UTI), bladder outlet obstruction, prolapsed urethral mass, etc[3]. An ureterocele related with the upper pole of the kidney is generally named as ectopic or pediatric type[1]. Dysplastic upper pole renal units in association with duplex system ureteroceles can also cause some management problems.

Hydronephrosis and ureteral dilatation are also evidences for the impairment of nephro-urinary system. Ureterocele diagnosis and thorough evaluation need a combination of radiological and nuclear scintigraphic methods. A sonogram and voiding cystourethrography are essential initial procedures for a child suspected of having ureteral anomaly[1,3,4].

Treatment failures can cause hypertension and end stage renal disease. There is still controversy and challenge on management protocols of ureterocele[3,4]. Today, endoscopic incision is an initial procedure for management. Even though it is the simplest and least invasive form of treatment particularly for single system ureteroceles, some urologists prefer ureterocele excision and ureteral reimplantation because of the risk of inducing reflux[6].

Reconstructive surgery is still a preferred method particularly for the duplex system ureteroceles and in the presence of vesicoureteral reflux[7]. Upper pole nephrectomy and ureterectomy is one of the treatment options for ureteroceles particularly in the absence of reflux; some excised specimens show only obstructive or inflammatory changes[2,5].

We planned to evaluate our patients with ureterocele diagnosis particularly by giving emphasis to these points of interest to discuss different management options.

Subjects and Methods

Data of the patients treated for ureterocele from the pediatric surgical departments of the two hospitals (İzmir Behcet Uz Children's Hospital and Celal Bayar University Hospital Manisa) during 1996-2009 were retrospectively evaluated. Inclusion criteria covered patients whose treatment was completed in our surgical departments. The patient records were reviewed regarding sex, age, antenatal diagnosis, symptoms related with ureterocele, pathological type and localization of the anomaly, radiological diagnosis, Dimercapto-succinic acid scinti-graphy, ureterocele-urinary system interaction, surgical treatment of ureteroceles and outcome.

Findings

Nineteen patients with ureterocele were treated during a 14 year period. There were 12 girls and 7 boys. The average age of the 14 patients older than 1 year was 5 years. Three patients with antenatal diagnosis were documented as 'bilateral hydronephrosis', 'unilateral hydro-nephrosis' and 'left renal cyst'. Symptoms and diagnostic reasons for the patients are summarized in Table 1. Ureterocele localizations

Table 1: Symptomatology and other diagnostic reasons for the patients with ureteroceles

First application complaints	Number of the patients
Evaluation for urinary tract infection	12
Nocturnal enuresis	1
Hydronephrosis (2 patients antenatally diagnosed)	3
Prenatal cyst	1
Posttraumatic hematuria	1
Incidentally diagnosed	1

Fig. 1: Four-year old girl presented with urinary tract infection and left hydronephrosis treated by endoscopic incision. **1a:** Huge bladder diverticular appearance of left ureterocele in VCUG (Left). **1b:** Ultrasonographic appearance of the ureterocele at the left side (Right).

were right in 9 patients, left in 9 patients and bilateral in 1 patient. Association of duplex systems was detected in 11 patients (6 at the same side, 3 bilateral and 2 contralateral).

Voiding-cystourethrography (VCUG): We performed VCUG in 14 patients and VUR was diagnosed in 4 (33%). One patient had Grade 1 and three patients had Grade 2 VUR. VCUG revealed ureterocele in four patients, and one patient was misdiagnosed as 'bladder diverticulum' (Fig. 1a). Four patients showed normal images in VCUG.

Ultrasonography (US): Ureterocele was defined by US showing cystic lesion in the urinary bladder in 13 out of 17 patients (Fig. 1b). Dilated distal ureters were revealed by US in 2 patients. Duplex systems were diagnosed in 5, and dilated urinary systems with varying degrees were detected in 15 patients. Ureterocele was incorrectly diagnosed with US as 'bladder diverticulum' in one patient.

Intravenous urography (IVU): IVU showed ureterocele in 8 of the 12 patients. Duplex systems were found in 5 patients. Nine patients had dilated urinary systems (Fig. 2). Remarkable upper pole dysfunction was shown in one patient by this method. The patient was treated by endoscopic incision.

Ureterocele-urinary system interaction: In general 15 out of 19 patients were ascertained as having varying degrees of dilated urinary systems - either unilateral or bilateral - depending on the radiological diagnostic methods.

Dimercaptosuccinic acid scintigraphy (DMSA): Ipsilaterally localized renal scarring and non-functioning upper pole images were taken in 7 of the 13 patients (Fig. 3a).

Computed tomography and *MAG_3 scintigraphy* were used in one of our patients for differential diagnosis of hydronephrotic mass and obstruction (Fig. 3b).

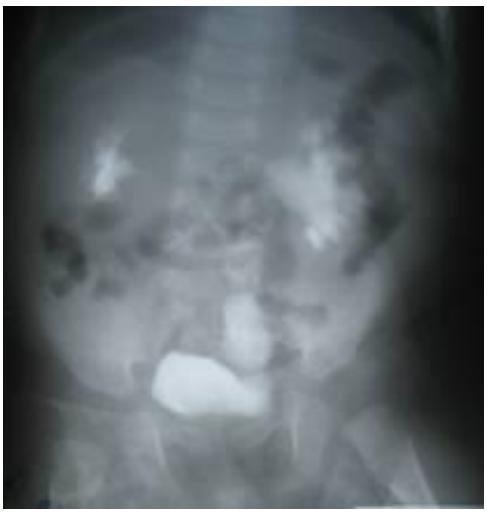

Fig. 2: Grade3-4 hydroureteronephrosis of a 1.5-year old boy with antenatal diagnosis of bilateral hydronephrosis in IVU.

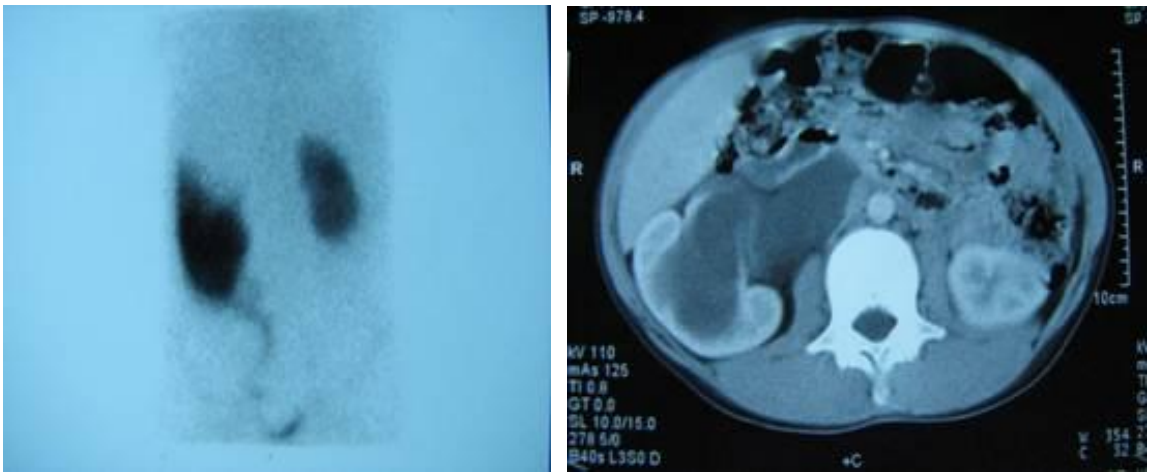

Fig. 3: Ten-year old boy presented with posttraumatic hematuria. Ureterocele excision and bilateral ureteral reimplantation was performed for right ureterocele and double collecting systems. **3a:** Wide hypoactive area, dilated and tortuous ureter at the right; hypoactive area at the lateral middle pole of the left kidney with differential functions of 65% right and 35% left in the DMSA scintigraphy (Left). **3b**: Grade 4 hydronephrosis and hydroureter in CT section (Right).

Cystoscopy was used for complementary diagnostic reasons in patients who underwent open surgery and for all patients who were endoscopically treated.

Surgery for ureteroceles: Surgical therapies that we performed in patients are summarized in Table 2.

Pathological evaluation: Tissue specimens of 11 patients were investigated. 'Chronic pyelo-nephritis' was diagnosed in resected upper pole kidneys of 3 patients and 'ureteritis' (in association with acute or chronic inflammation) detected in the samples of excised ureterocele tissues of the other 8 patients.

Patients' surveillance: One patient died from a probable neurological intracranial lesion. One patient was followed and treated for arterial hypertension for years. One patient had refractory UTIs in the follow up period. We saw postoperative urinary tract infection in three (two boys, one girl) of the six endoscopically treated patients. The others were free of infection. Postoperative reflux was seen in two patients, one of them had been endoscopically

Table 2: Type of the surgical procedures applied

I. Upper Pole Nephrectomy (3)
Upper pole nephrectomy * only (1)
Upper pole nephrectomy *+ ureterocele unroofing (1)β
Upper pole nephrectomy *+ double sided ureteral reimplantation+ ureterocele excision (1)δ
II. Bladder Level Surgery
A. Open Surgery (12) Ø
Ureterocele excision # (3)
Ureterocele excision # + double sided ureteral reimplantation (7)
Ureterocele excision # + same side ureteral reimplantation (1)
Ureterocele unroofing (1)
B. Endoscopic treatment (Ureterocele incision) (7)α

* Combined with ureterectomy/ β: One stage operation/ δ: Two staged operation/ Ø: Eight duplex systems + four single systems/ # Ureteroceles were totally excised, defective bladder muscles were repaired and ureteral tapering was not performed/ α: All patients but one had single system ureteroceles and bladder level reconstruction was needed for one patient subsequently)

treated. One patient was followed for micturating troubles with bladder dysfunction that became symptomatic postoperatively and lost renal function. None of the patients showed recurrence of the disease except one patient who was recently learned to be reoperated in another center for ureterocele.

Discussion

Ratio of gender distribution in our series was 1:7 (Female/Male) and the average age of our patients was calculated as 5 years. According to literature, 90 % of the patients with ureterocele are diagnosed before the age of 3 years[8]. Though there is an increasing number of antenatally diagnosed patients with urological anomalies, 'toddler age' is still the most common period that we treat the patients with ureterocele[4,5]. Boys are known as more problematic during the newborn or infancy periods, but symptomatic female infants have been treated who also showed management difficulties[1].

Most of the patients with ureterocele are classically diagnosed during the investigation for UTI, asymptomatic hydronephrosis and loin mass[3]. Although the age of diagnosis is decreasing, UTI is still the most common clinical presentation of ureterocele in 50% of the patients promoting physician to make the thorough evaluation of the urinary system[1,8].

UTI was a remarkable finding in our patients with a rate of 63%. Depending on the pathological findings such as ureteritis and inflammatory changes detected in the excised tissue specimens, it is suggested that UTI helps destruction of urinary tissues as these findings are rarely primary when there is any associated abnormality such as ureterocele, megaloureter, etc [9].

Duplex system association is a common finding for ureteroceles. Sixty percent of our patients had this combination. We had relatively more patients (seven) with single system ureteroceles. Though single system ureteroceles are known as relatively simple abnormalities, they also can show management challenges and damage nephrourinary system like we encountered in our patients.

A combination of diagnostic methods is used for preoperative evaluation of ureteroceles. Whole nephrourinary system could have already been negatively affected at the time of diagnosis. Ultrasonography is an easy to perform, non-invasive and probably the best imaging modality for making the diagnosis[2]. We found it as the most successful procedure (76% in the series) in ureterocele diagnosis and for the evaluation of hydro-ureteronephrosis. It is also the recommended screening method after the first urinary tract infection[1]. Sonography scan is able to catch the lesions which are not obvious on VCUG[3].

VCUG is used for ureterocele diagnosis and detection of VUR. Reflux can occur into the ipsilateral lower pole in almost half of the patients, but contralateral system is also affected with a rate of 25%[1-3,5]. Although lower grades (Grade 1-2) of VUR were detected in our patients, we strongly recommend therapy for reflux in complicating urinary tract infections. In our opinion, those refluxes in association with ureteroceles should not be managed as if VUR were the only underlying pathology causing UTI. VCUG is also beneficial for following up patients with preoperative VUR diagnosis or to detect newly forming refluxes after endoscopic interventions[4].

We believe that IVU has not completed its role as a diagnostic modality. Although IVU is not the currently preferred method, it helps to determine the management protocol of the surgical procedure by showing ureterocele (in 66% of our patients), displaying anatomical pathology characteristics and the non-functioning upper poles[3].

DMSA scintigraphy should be undertaken routinely to assess the distribution of function in the duplex kidney and for detecting and follow up of scarred tissue and non-functioning upper poles[1,4,6,10]. We found renal scarring in nearly 55% of our patients which is a point of attraction during the management of ureteroceles. Scarring indicates parenchymal disruption by ways of dysplastic changes, UTI and VUR.

In current era of endoscopy, upper pole nephrectomy is not a preliminary technique for the treatment of ureteroceles. We performed this surgical method at the early times of the study period. The upper renal units related with ureteroceles show histopathological changes.

Currently we agree with some authors who suggest that upper poles with lower function could be left in their places after bladder level treatment (either endoscopically or surgically) of the ureterocele as these lesions were not progressive or reversible and inflammatory tubulointerstitial nephropathy was the most common pathology encountered in the dysplastic kidneys[5,11-14]. Chronic inflammatory findings (Chronic pyelonephritis) could be the result of or related with recurrent or chronic UTIs. There is no one report showing the development of malignancy based on this remnant tissue.

As we experienced, attention should be paid to diverticular misdiagnosis because a different type of management protocol could be needed.

With voiding, the ureterocele prolapses through the detrusor and can mimic a bladder diverticulum. Therefore, ureterocele diagnosis should be confirmed by other diagnostic tools[1,2,5] (Figure 1a).

Ureterocele and associated pathologies cause dilatation of the pelvi-calyceal systems and ureter by different ways, such as obstruction, VUR or primary dysmorphism (Figures 2, 3b)[1,6]. Ureterocele is an important pathology causing dilated systems at the time of diagnosis as we detected in 14 (84%) of our patients.

Dilated urinary systems may play a role in insisting UTIs during the postendoscopic intervention period by causing improper peristaltic activity and deficiency of urine propulsion.

We have been doing endoscopic surgery for the last eight years. This simple and safe procedure is recommended as the first line treatment of complete duplex system with intravesical ureterocele, particularly for newborns or septic states before the age of one year[15,16]. Although ureterocele incision is a good alternative to other surgical methods, all patients need to be followed up for VUR, UTI and hypertension[17]. Individualization in the therapy is still important; there is not only one surgical method suitable for every type of the anomaly and challenges on this subject still continue [3,4].

In our opinion bladder level surgery conserves its importance. Ectopic ureteroceles in association with VUR particularly are best treated by excision of ureterocele and ureteral reimplantation[14,18]. Although low grade (Grade 1-2) refluxes tend to resolve spontaneously in time, most are symptomatic when associated with ureteroceles[3]. For many patients the real treatment free status most often requires ureterocele excision. A staged approach with initial trans-urethral incision, followed by excision and reimplantation can be radical for treating much of the patients[13]. The type of lower urinary tract reconstruction -- whether total or partial excision of ureterocele -- does not seem to affect results[19]. We made total excision and repaired the defective bladder muscle in patients whom we performed ureterocele excision as bladder level surgery (Table 2). We didn't perform ureteral tapering. Similarly dilated ureters are always left untouched in endoscopic ureterocele interventions without complications.

Regular follow-up of the patients in outpatient departments for longer periods cannot be economically efficient. The more important issue is that patients can suffer from morbidity when they fail to comply with doing regular visits. Beyond any doubt, reflux nephropathy is an important and premium cause of chronic renal failure and renal transplantation in many countries.

Individualization in therapy due to different pathological and clinical characteristics of the disease is important and often there is no single solution for the patients; therefore, family opinion should be added to the management protocol[1,14,20].

Conclusion

We are still making the diagnosis of ureteroceles in relatively older ages, more often during investigation for UTI and VUR. Although UTI

caused some problems in the postoperative period, and time was limited to our follow-up periods, none of the patients needed a second operation because of recurrence of the disease or from other complications after radical surgery except one patient mentioned above.

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Conflict of Interest: None

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